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PINEALOMA WITH SIMMONDS' SYNDROME. REPORT OF A CASE

by

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It has been well known since Berblinger (1925) that pinealomas usually invade and destroy adjacent structures and penetrate the third ventricle, and occasionally they may spread to the ependyma or subarachnoid space by implantations through the cerebrospinal fluid. A number of peculiar cases have been reported, in which the whole ventricular wall is replaced by the layer of metastatic pinealoma tissue. In such cases various clinical manifestations were present. However, only a few cases have been reported in the literature, in which pinealomas were associated with Simmonds' syndrome. In the present case of a pinealoma, the patient presented the signs and symptoms simulating Simmonds' syndrome without those of increased intracranial pressure.

CASE REPORT

K. T., male, 17 years of age, was admitted to the First Medical Clinic of our Hospital on Nov. 18, 1951, complaining of lassitude and loss of appetite.

The patient had been in excellent health until August 1950. In the beginning of Sept. of the same year, he began to complain of lassitute and loss of appetite. He consulted various physicians, who, because of the diminution of his body weight, gave him hypophysial extracts. But, there was no improvement. In addition, about the end of 1950, he began to complain also of polyuria and dizziness, and became unusually nervous. His body weight gradually diminished from 39kg to 29kg in about one year before admission.

Examination. On admission he was very weak and emaciated. The skin was delicate and dry. Pubic and axillary hair was scanty. However, neurologically there were no abnormalities. Blood-pressure was 88/60mm. Hg. and body temperature normal. Basal metabolic rate was-46%. Pulse rate was 60 per minute. The rate of excretion of 17-ketosteroids in the urine was 6.6mg per day. Daily amount of urine was 2000-3000cc.

The diagnosis was an endocrine disorder, presumably Simmonds' disease, and he was given various kinds of hormones.

Several months later, about March 1952 visual disturbance appeared and grew worse rapidly, until he became blind with the right eye and 0.1 with the left eye at the end of April 1952. In addition, the paralysis of upward conjugate movement and some nystagmoid jerks of the eyes on the looking to the left were noted.

Then he was referred to our clinic on June 5, 1952 for neurosurgical treatment.

There was an outstanding emaciation. However, he complained neither of headache, nor vomiting. There was no edema of the optic discs. Roentgenogram of the skull showed no abnormalities of the sella turcica. The pressure of the spinal fluid was 220mm of fluid in the recumbent posture. The fluid was otherwise normal. By the clinical impression the disease seemed not to be a brain tumor, but an optochiasmatic arachnoiditis. For the relief of his rapidly diminishing vision, an immediate operation was advised.

Operation. A transfrontal osteoplastic operation was performed on the right side. Without any great difficulty the optochiasmatic region was reached, but the tumor was not found. The optic nerves were found to be surrounded by tortous dilated veins which were seen like racemous angioma. Moreover, the abnormalities in shape and course of the cerebral meningeal artery and its branches were found. The wound was carefully closed. The visual acuity was not improved. From the end of August 1952 he became lethargic and slept most of time. Besides neurogenic hyperthermia of 39.5°~40.5°C appeared and persisted until his death on Nov. 7, 1952.

Postmortem Examination. Unfortunately the autopsy was limited to the head. It revealed a large soft tumor, as large as a hen's egg, best seen in the midline sagittal section of the brain, extending from the pineal body to the hypothalamus. The tumor was irregular in shape and the epithalamic portion of the tumor was smaller than the hypothalamic portion. The limit of the tumor in the third ventricle was diffuse (Fig. 1).

The microscopic structure was characteristic of a pinealoma (Figs. 2 and 3). The tumor was composed essentially of masses of large epitheloid cells separated by a network of reticular connective tissue containing numerous lymphoid cells. The epitheloid cells had large spherical nuclei, each of which contained one or two nucleoli and very little chromatin. (Fig. 2 shows the epithalamic portion of the tumor and Fig. 3 the hypothalamic portion.)

In the stalk (Fig. 4) and in the posterior and intermediate lobes of the hypophysis (Fig. 5), the tumor cells were exclusively lymphoid cells which were diffusely distributed, forming lymphoid follicles here and there.

The anterior lobe of the hypophysis (Fig. 6) was infiltrated by the tumor cells partially, and the nearer to the posterior lobe the greater was the infiltration, accompanied with many degenerative features of chromophobe and chromophil cells.

The number of various cells of the anterior lobe was estimated by the method of Rasmussen (1929). The distinguished diminution of chromophil cells, especially of eosinophil cells was seen (Table 1).

DISCUSSION

Our case is a pinealoma, in which the implantation metastasis in the hypothalamus was more marked than the primary pineal tumor, and the metastatic tumor cells invaded also the posterior part of the hypophysis, although it was not certain that the portion of the tumor in the pineal region was primary and that the

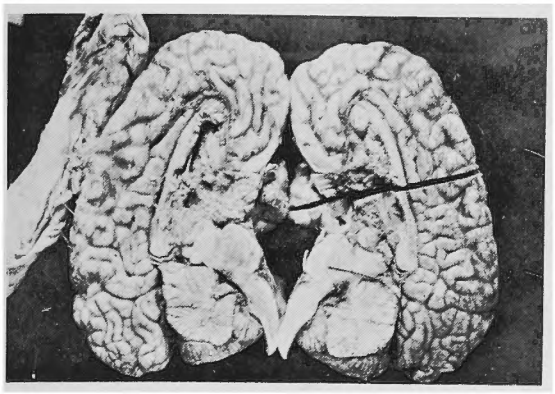

Fig 1

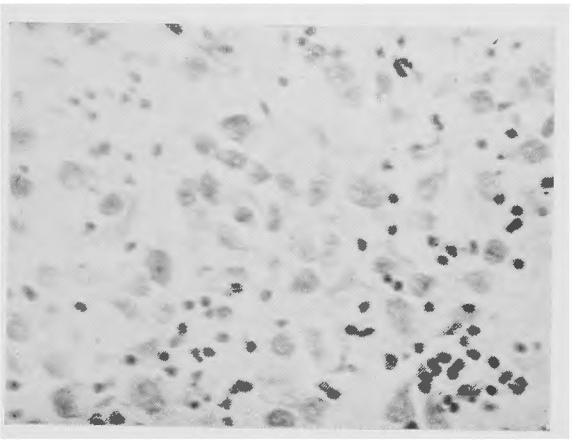

Fig 2

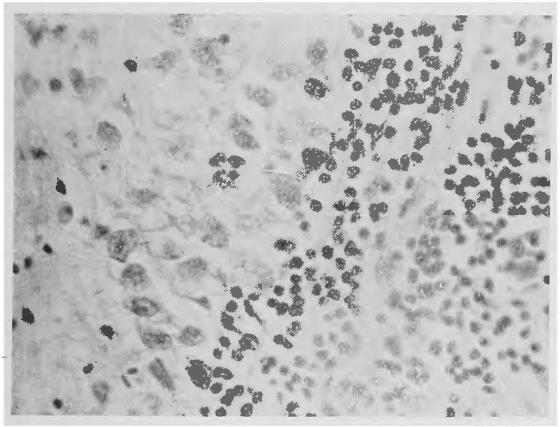

Fig 3

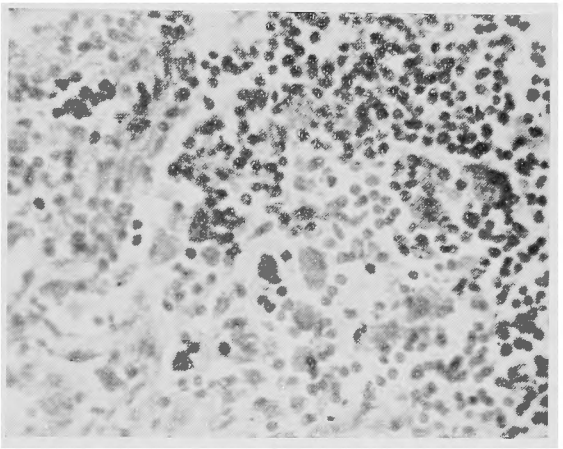

Fig 4

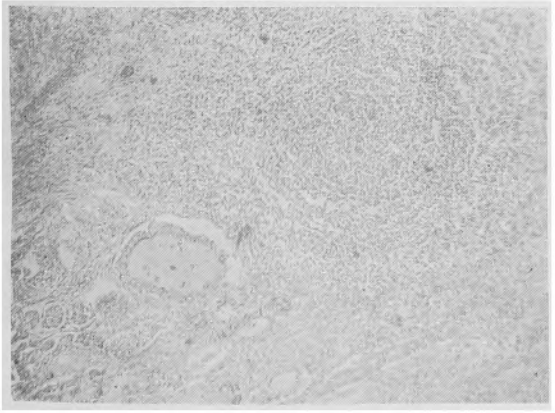

Fig 5

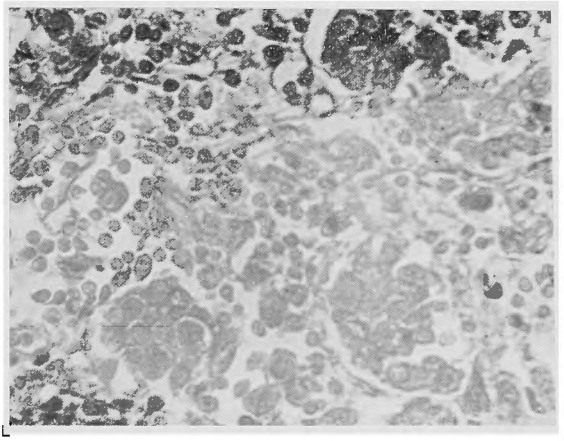

Fig 6

Table 1. Proportion of cell types of the anterior lobe of the hypophysis

	chromophobe cells	eosinophil cells	basophil cells
Rasmussen (normal Europeans)	34-66% (52.2%)	23-60% (36.8%)	4-27% (10.9%)
Inamoto (normal Japanese)	38.6-62.6% (52.7%)	22.3-45.5% (34.0%)	3.6-25.7% (13.3%)
This Case	79.7%	13.5%	6.8%

hypothalamic portion was secondary. As for the occurrence of ectopic pinealomas, it has been generally accepted since R.P. Mackay (1939) that the pineal body is nothing but a highly differentiated ventricular ependymal tissue, and accordingly there is a possibility of arising of a pinealoma from any part of the ventricular ependyma. However, the disseminated metastases of pinealoma in the subarachnoid space or in the ventricular wall by the flow of the cerebrospinal fluid are more widely known, as reported by Berblinger (1925) and others. Therefore it is more easily acceptable that the primary tumor in our case originated in the pineal region.

However, it is of interest to note that the patient presented the signs and symptoms simulating Simmonds' syndrome without those of increased intracranial pressure. One may be puzzled over why such symptoms should have developed in this case. As regards the lack of intracranial hypertension at least in the early stage, the high malignancy of the tumor may be supposed, i. e. the tendency of the tumor to form easily dissemination metastases over the ventricular wall before the primary pineal tumor becomes large enough to obstruct the aqueduct. It is evident in Fig. 1 that the epithalamic portion of the tumor is rather small as compared with the large hypothalamic portion. In this connection the fact may be of some significance that in our case the tumor cells in the infundibulum as well as in the intermediate part of the hypophysis were exclusively lymphoid cells which were diffusely distributed.

As for the Simmonds' syndrome it is to be noted that the greater destruction was found in the stalk and the posterior portion of the pituitary than in the anterior portion. In considering that the destruction was the most striking in the hypothalamus, the syndrome appearing in this case should be considered to be not of anterior-pituitary origin but of hypothalamic origin.

SUMMARY

A case is reported of a pinealoma with Simmonds' syndrome. The primary tumor in the pineal region is rather small and a larger metastatic tumor is found in the hypothalamus, invading the infundibulum and the posterior lobe of the hypophysis.

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